Diffuse plane normolipaemic xanthomatosis as a manifestation of monoclonal gammopathy

Morsink, Linde M.; Nijhof, Inger S.

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A 48-year-old male presented with progressive yellow colourisation of his skin in the axillary and inguinal regions (images). His blood count, renal function tests, calcium, cholesterol and triglyceride levels were normal, but serum protein electrophoresis showed an immunoglobulin G kappa monoclonal protein. Complement C4 levels were undetectable, with normal C3 and low C1 esterase inhibitor levels. No skeletal lesions were detected on computed tomography and bone marrow contained 1% monoclonal plasma cells. Skin biopsy showed no amyloidosis, but aggregates of foamy macrophages intradermally, compatible with diffuse plane normolipaemic xanthomatosis, associated with a monoclonal gammopathy. Pathogenesis involves the monoclonal protein forming immune complexes with lipoproteins in the skin with subsequent precipitation, followed by phagocytosis by macrophages; there is simultaneous activation of the classic complement pathway resulting in low levels of C4 and C1 esterase inhibitor (Szalat *et al*, 2011). The patient has received no treatment so far and is currently clinically well.

Linde M. Morsink[1] **and Inger S. Nijhof**[2]

[1]*Department of Haematology, University Medical Centre Groningen, Groningen, the Netherlands and* [2]*Department of Haematology, Amsterdam University Medical Centre, Amsterdam, the Netherlands.*

E-mail: linde.morsink@gmail.com

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